Aortic translocation for the management of transposition of the great arteries with a ventricular septal defect, pulmonary stenosis, and hypoplasia of the right ventricle

Victor O. Morell*, Peter D. Wearden

Section of Pediatric Cardiothoracic Surgery of the Heart, Lung and Esophageal Surgical Institute, University of Pittsburgh Medical School, Children’s Hospital of Pittsburgh, Pittsburgh, PA, United States

Received 19 September 2006; received in revised form 13 November 2006; accepted 20 November 2006; Available online 9 January 2007

Abstract

A 2-month-old patient with transposition of the great arteries, a ventricular septal defect, pulmonary stenosis, and severe hypoplasia of the right ventricle successfully underwent a biventricular repair utilizing the aortic translocation technique. Advantages of this surgical repair over the Rastelli procedure in the management of this complex congenital heart lesion are discussed.

© 2007 European Association for Cardio-Thoracic Surgery. Published by Elsevier B.V. All rights reserved.

Keywords: Aortic translocation; Rastelli repair; Transposition of the great arteries

1. Introduction

Right ventricular hypoplasia in the setting of transposition of the great arteries (TGA) with a ventricular septal defect (VSD) and pulmonary stenosis (PS) represents a major anatomic obstacle in achieving a biventricular repair. The Rastelli procedure, although a surgical option, is not ideal because the creation of an intraventricular tunnel will further reduce the size of the small right ventricular cavity. The aortic translocation repair provides a sound anatomic repair, preserving the right ventricular volume. We describe the case of a 2-month-old infant with TGA, VSD, PS, and a hypoplastic right ventricle who successfully underwent a biventricular repair utilizing the aortic translocation technique.

2. Case report

A 2-month-old male infant born with d-transposition of the great arteries with a VSD and PS was referred for surgical repair at our institution. His room air saturations were in the mid to low 70s with no previous palliative procedures. The preoperative echocardiogram revealed the presence of moderate valvar and subvalvar pulmonary stenosis (Fig. 1A); the pulmonary valve annulus measured 0.69 mm in diameter and the valve was bicuspid. There was moderate right ventricular hypoplasia (Fig. 1B), with a tricuspid valve annulus Z-score of −2.5.

Initially, the patient underwent an aortic translocation procedure that included an en-block transfer of the aortic root and coronary arteries, with a Lecompte maneuver, VSD closure, and a direct right ventricle to pulmonary artery anastomosis. Once off cardiopulmonary bypass (CPB), there were ST changes in the electrocardiogram (EKG) suggestive of coronary ischemia. We believed that the ischemic changes were most likely secondary to torsion of the proximal coronary arteries after translocating the aortic root. Therefore, under cardiac arrest, the coronary arteries were harvested and reimplanted in a more anterior position in the aorta. The patient was then weaned from CPB with normal biventricular function and a normal EKG. The patent foramen ovale was not closed in order to allow for right-to-left shunting at the atrial level.

His postoperative course was complicated by postoperative bleeding requiring mediastinal exploration on the night of surgery. On postoperative day #3, approximately 12 h after extubation, the patient developed respiratory compromise precipitating a cardiac arrest. Extra-corporeal membrane oxygenation (ECMO) support was implemented during resuscitation and was continued for a 24-h period after which the patient was weaned without difficulty. He developed post-ECMO seizures that were controlled with medication, but had no significant neurological deficits. At the time of discharge,
his room air saturation was in the low 90s, secondary to a right to left shunt at the atrial level. The discharge echocardiogram revealed normal biventricular function with normal flow across both the ventricular outflow tracts.

3. Discussion

Transposition of the great arteries with a ventricular septal defect and pulmonary stenosis represents approximately 25% of all transpositions. Hypoplasia of the right ventricle in this subset of patients is rare but, when present, significantly impacts on the surgical options for biventricular repair. Preservation of the right ventricular volume is of major importance.

Since it was first described in 1969, the Rastelli procedure has become the most common surgical technique utilized in the management of this lesion. The main technical component of this repair consists of the creation of an intraventricular tunnel connecting the posterior left ventricle to the anterior aorta. Anatomically, this tunnel sits in the outflow portion of the right ventricle, therefore reducing the volume of the right ventricular cavity (Fig. 2). Obviously, patients with a hypoplastic right ventricle are not good candidates for this repair.

In 1980, Bex et al. [1] reported a new surgical technique for the management of transposition of the great arteries. They described moving the aortic root together with the coronary arteries to the pulmonary position as a way to provide a true ‘anatomic correction’ for TGA. The procedure was perfected in anatomical specimens before performing it successfully in a 3-year-old girl with TGA and pulmonary stenosis. In 1984, Nikaidoh [2] described and popularized the concept of aortic translocation for the management of TGA with a ventricular septal defect and pulmonary stenosis.

The technique of aortic translocation [3] involves the harvesting of the anteriorly placed aortic root from the right ventricle with division of the proximal pulmonary artery and the conal septum. The aortic root is then translocated to the pulmonary position (posterior) so that it will lie on top of the left ventricle, avoiding creation of an intraventricular tunnel. This technical point represents a major surgical advantage for patients with a hypoplastic RV in which we are trying to preserve as much right ventricular volume as possible.

Fig. 1. (A) Echocardiograph sub-costal view showed left ventricular outflow tract obstruction (LVOTO) secondary to posterior deviation of the conal septum. (B) Echocardiograph 4-chamber view showed moderate right ventricular hypoplasia and a small tricuspid valve. (LA, left atrium; LV, left ventricle; PA, pulmonary artery; RV, right ventricle; RA, right atrium; VSD, ventricular septal defect).

Fig. 2. (A) Lateral view of the heart with transposition of the great arteries, ventricular septal defect, pulmonary stenosis, and a hypoplastic right ventricle after a Rastelli procedure. Note the reduction of right ventricular volume secondary to the creation of an intraventricular tunnel. (B) Lateral view of the same heart after the aortic translocation procedure. This surgical technique results in better preservation of the right ventricular volume and better alignment of both ventricular outflow tracts. (Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RV, right ventricle).
Certainly, as in the case of patients with pulmonary atresia with an intact ventricular septum, there is a degree of right ventricular hypoplasia that precludes the establishment of an adequate biventricular circulation. It is well documented [4,5] that the tricuspid valve size plays a major role in our ability to achieve a complete repair in patients with a hypoplastic right ventricle. Our patient’s tricuspid valve Z-score of greater than −3 was felt to be sufficient for a successful biventricular repair. Also, the option of a one and a half ventricle repair was considered.

4. Conclusion

We believe that the aortic translocation repair is superior to the Rastelli procedure in the preservation of right ventricular volume, and therefore, should be the procedure of choice for the management of patients with transposition of the great arteries with a ventricular septal defect, pulmonary stenosis, and a hypoplastic right ventricle.

References